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Pediatric Cardiology

Factors Associated With Outcomes of Persistent Truncus Arteriosus

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OBJECTIVES The purpose of this study was to identify trends and factors associated with outcomes of persistent truncus arteriosus (PTA).

BACKGROUND Although there have been significant improvements, PTA continues to be associated with significant morbidity and mortality.

METHODS We undertook a review of all consecutive cases of PTA (n = 205) presenting at our institution from 1953 to 1997. Data were collected regarding demographics, anatomy, management (surgical palliation and repair) and outcomes (mortality and reoperation).

RESULTS Significant trends ($p \leq 0.001$) related to groups defined by year of birth were as follows: number of cases (1953–1967, n = 13; 1968–1977, n = 42; 1978–1987, n = 69; 1988–1997, n = 81), median age at first assessment (8 months, 42 days, 7 days and 2 days, respectively), proportion who did not have any surgery (58%, 27%, 22% and 11%), proportion who had an initial palliative procedure (25%, 37%, 6% and 2%), proportion who underwent PTA repair (31%, 59%, 72% and 88%), median age at PTA repair (11.2 years, 1.1 years, 1.6 months and 12 days) and proportion dying before hospital discharge after repair (50%, 63%, 56% and 41%). Since 1995, mortality before hospital discharge after repair has further decreased to 2/11 (18%). Increasing time to initial conduit replacement in hospital survivors was significantly related to larger sized conduit at repair ($p = 0.02$) and use of pulmonary homografts (vs. aortic homografts or xenografts; $p = 0.002$). Interventional catheterization to address conduit obstructions significantly increased conduit longevity.

CONCLUSIONS Significant improvements in PTA outcomes are evident with trends toward earlier age at assessment and complete repair. (J Am Coll Cardiol 1999;34:545–53) © 1999 by the American College of Cardiology

Persistent truncus arteriosus (PTA) is an uncommon congenital cardiac malformation accounting for approximately 1.2% of all congenital heart malformations (1). Features of PTA include a single great artery arising from the base of the heart, that supplies systemic, coronary and pulmonary blood flow, together with a ventricular septal defect. Both characteristics are believed to result from failure of proper conotruncal septation. The two main classification systems used to describe the anatomy of PTA are those of Collett & Edwards (1949) and Van Praagh (1965) (2). The natural history of children born with PTA is such that up to 80% or more die within the first year of life without surgical intervention (1,3,4,5). The first successful surgical repair of PTA was performed by McGoon, Rastelli and Ongley (6) in 1967. They used an aortic homograft including the aortic valve to establish continuity from the right ventricle (RV) to the pulmonary artery (PA). Since then, various means of reconstruction have been developed, and controversies remain concerning which method is the best to use and at what age surgery should optimally be performed. The main long-term management issues relate to conduit stenosis and truncal valve incompetence and stenosis. The necessary reoperations to address these complications are associated with further risk of mortality and morbidity. Risk factors and outcomes related to this lesion have been incompletely defined. We therefore sought to determine the changing patterns of mortality and morbidity related to PTA at a single institution and to define potential factors associated with outcomes related to management.

METHODS

Study population. All patients with a diagnosis of PTA were identified in the Cardiology Database of the Division of Cardiology, The Hospital for Sick Children, and were included in the present study. The diagnosis was verified in the medical record for all patients, and patients who were miscoded and did not have PTA were excluded.

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Abbreviations and Acronyms
PA = pulmonary artery
PTA = persistent truncus arteriosus
RV = right ventricle

Measurements. The cardiology and hospital records for all patients were reviewed. Data collected included demographics, anatomic diagnoses, status at presentation, surgeries, interventional catheterization procedures and follow-up status.

Data analysis. Data are described as frequencies, medians with ranges and means with standard deviations. Where there is missing data, the available values are given. Changes in characteristics, management and outcomes over time were sought by creating four groups based on birth date: 1953–1967, 1968–1977, 1978–1987 and 1988–1997. Differences between these groups were sought using Mantel Haentzel chi-square tests, Kruskal-Wallis analysis of variance and analysis of variance. Associations between the number of PTA valve cusps and presence, and severity of stenosis and regurgitation were sought with Mantel-Haentzel chi-square tests. Severity of stenosis was related to severity of regurgitation with gamma coefficient. Factors associated with hospital mortality after PTA repair were sought with Fisher exact tests, chi-square tests, Kruskal-Wallis analysis of variance and Student *t* tests. Kaplan-Meier estimates of freedom from initial RV to PA conduit replacement were plotted, and factors associated with decreased time to initial conduit replacement were tested with log rank and Wilcoxon tests. A $p < 0.05$ was set as the level of statistical significance.

RESULTS

Patient Characteristics

Demographics. From the Cardiology Database, 205 patients were identified and verified with the diagnosis of PTA, and date of birth ranged from January 1, 1953 to December 31, 1997. Cases were grouped into birth cohorts as follows: 1953–1967, n = 13; 1968–1977, n = 42; 1978–1987, n = 69 and 1988–1997, n = 81. There were 76 males and 129 females.

Presentation. The median age at presentation (n = 202) was 4 days with a range from birth to 5.8 years. Median age of participants at presentation became progressively younger with subsequent birth cohorts ($p < 0.001$) as follows: 1953–1967, 92 days (n = 13); 1968–1977, 17 days (n = 41); 1978–1987, 2 days (n = 68) and 1988–1997, 1 day (n = 80). Clinical signs at presentation (n = 201) were congestive heart failure in 117 patients (58%), cyanosis in 89 (44%), respiratory infection or distress in 33 (16%), failure to thrive in 7 (3%), cardiac arrhythmias in 3 (1%) and clubbing of fingers and toes in two patients (1%). One patient presented in a moribund state. The methods used to establish the diagnosis (n = 200) consisted of echocardiography alone in 103 patients (51%), cardiac catheterization alone in 63 (31%), both echocardiography and catheterization in 29 (15%), echocardiography with magnetic resonance imaging in 2 (1%) and at postmortem examination in 3 patients (2%).

The median age at diagnosis (n = 202) was 7 days with a range from 22 days before birth at fetal echocardiography to 15 years. Median age at diagnosis became progressively younger with subsequent birth cohorts ($p < 0.001$) as follows: 1953–1967, 8 months (n = 13); 1968–1977, 42 days (n = 41); 1978–1987, 7 days (n = 68) and 1988–1997, 2 days (n = 80).

Cardiac anatomy. The specific PTA anatomy of the patients (n = 202) was as follows: single pulmonary trunk and ascending aorta arising from the PTA in 140 patients (69%), right and left PAs arising from separate orifices close together from the dorsal wall of the PTA in 41 (20%), one or both PAs arising from the separate orifices at the lateral aspects of the PTA in 14 (7%), the branch PAs immediately adjacent to one another with a common orifice but no common trunk following attachment from the PTA in 4 (2%), no true branch PAs arising from the PTA (Collett and Edwards Type 4) in 2 (1%) and in one patient, the right PA arose from the posterior wall of the PTA with the left PA arising from a main PA from the PTA (<1%). Other associated cardiovascular anomalies are described in Table 1. The reported prevalence of interatrial communications reflects only those defects noted at echocardiography, cardiac catheterization, surgery or autopsy and may be an underestimation of defects that may not have been adequately imaged or that closed spontaneously.

The number of PTA valve leaflets (n = 156) were two in 11 patients (7%), three in 85 (54%), four in 58 (37%), five in 1 (1%) and six in 1 patient (1%). Persistent truncus arteriosus valve regurgitation was described in 123 (69%) of 178 patients and was graded subjectively at the time of either echocardiography or cardiac catheterization in 115 patients as mild in 88 (77%), moderate in 22 (19%) and severe in 5 patients (4%). Persistent truncus arteriosus valve stenosis was described in 75 (56%) of 135 patients and was graded subjectively in 72 patients as mild in 49 (68%), moderate in 11 (15%) and severe in 12 patients (17%).

Persistent truncus arteriosus valve abnormalities significantly impacted on the incidence of both valve stenosis and regurgitation. Moderate to severe valve stenosis was more likely in patients with two valve cusps (2 of 9; 22%) or four or more valve cusps (14 of 44; 32%) than it was in those with three valve cusps (5 of 65; 8%; $p = 0.005$). Moderate to severe valve regurgitation was more likely in patients with four or more valve cusps (17 of 57; 30%) than it was in those with two valve cusps (0 of 9; 0%) or three valve cusps (8 of 73; 11%; $p = 0.007$). The degree of valve stenosis was

Table 1. Associated Cardiovascular Anomalies (N = 202)

Anomaly	Patients	
	No.	%
Atrial septal defect/PFO	125	62
Right aortic arch	60	30
Patent ductus arteriosus	37	18
Stenotic or hypoplastic pulmonary arteries	32	16
Interrupted aortic arch	28	14
Aberrant right or left subclavian artery	12	6
Persistent left superior vena cava	12	6
Coronary artery anomaly	11	5.4
Coarctation of the aorta	3	1.5
Anomalous pulmonary venous drainage	3	1.5
Hypoplasia of the left lung	3	1.5
Anomalous origin of subclavian arteries	2	1
Single ventricle	1	0.5
Absent R-AV connection	1	0.5
Hypoplastic RV	1	0.5
Aneurysm of septum primum	1	0.5
AVSD	1	0.5
Tubular hypoplasia of aortic arch	1	0.5
PTA committed to LV only	1	0.5
Upstairs/downstairs ventricular relationship	1	0.5
Stenosis proximal LSCA	1	0.5
Azygous continuation of IVC	1	0.5

AVSD = atrioventricular septal defect; IVC = inferior vena cava; LSCA = left subclavian artery; LV = left ventricle; PFO = patent foramen ovale; PTA = persistent truncus arteriosus; R-AV = right atrioventricular; RV = right ventricle.

significantly correlated with the degree of valve regurgitation (gamma 0.526; error 0.099).

Noncardiac abnormalities. Noncardiac abnormalities were described in 81 patients and included DiGeorge syndrome (n = 22), dysmorphic features (n = 18), neonatal seizures (n = 10), cleft palate (n = 6), renal abnormalities (n = 6), bilateral choanal atresia (n = 3), cardiofacial syndrome (n = 3), esophageal atresia (n = 2), tracheoesophageal fistula (n = 1), Hemophilia A (n = 1), chest wall anomalies (n = 1), omphalocele (n = 1), quadriplegia (n = 1), absent corpus callosum (n = 1), Noonan syndrome (n = 1) and Potter syndrome (n = 1).

Management and Outcomes

The management and survivorship for the patients is shown in Figure 1. The management and status of three patients is unknown.

Presurgical outcomes. No surgery was performed in 42 (21%) patients; 31 (74%) of these patients subsequently died at a median age of 11 days (range: birth to 19.4 years), with 27 patients dying before 6 months of age. The median interval from diagnosis to death in these patients was 3 days (range: <1 day to 18.9 years), with one patient having been diagnosed at autopsy. With subsequent birth cohorts the proportion of patients who did not have any surgery decreased significantly as follows: 1953–1967, 7/12 (58%); 1968–1977, 11/41 (27%); 1978–1987, 15/69 (22%) and 1988–1997, 9/80 (11%; p = 0.001). Cause of death in most was due to metabolic acidosis with myocardial dysfunction, arrhythmia and cardiac arrest, with organ failure in other systems. Patients who died before any surgery were significantly more likely to have moderate to severe PTA valve stenosis at diagnosis (11/21; 52%) than the remaining patients (12/114, 11%; p = 0.001), but there was no significant difference in the incidence of moderate to severe valve regurgitation (7/28, 25%; vs. 20/150, 13%, respectively; p = 0.12). In addition, there were 11 patients who had no surgery who were last known to be alive at a median age of 5.0 years (range, 2 days to 18.1 years), and at a median interval from diagnosis of 4.7 years (range, 2 days to 17.8 years).

Palliative surgical procedures. Palliative surgical procedures were performed initially in 23/199 (12%) patients, at a median age of 2.5 months (range, 2 days to 2.5 years) and at a median interval from diagnosis of 18 days (range, <1 day to 1.3 years). Palliative procedures included pulmonary artery banding in 21 patients (in conjunction with repair of interrupted aortic arch in 2 patients) and placement of a systemic to pulmonary arterial shunt in 2 patients (in conjunction with repair of interrupted aortic arch in 1 patient). The proportion of patients whose initial surgery was a palliative procedure decreased significantly with subsequent birth cohorts as follows: 1953–1967, 3/12 (25%); 1968–1977, 14/38 (37%); 1978–1987, 4/68 (6%) and 1988–1997, 2/81 (2%); p = 0.001. In addition, there were significant trends with subsequent birth cohorts towards a younger age at surgery and a shorter interval since diagnosis in those patients who underwent palliative procedures. There were eight deaths (35%) in these patients, at a median interval of 1.5 days after surgery (range, <1 day to 12 days). Patients who died versus those who survived after palliative surgery were significantly younger at surgery (median age 11.5 days vs. 5.0 months, respectively; p < 0.05) with a trend toward a shorter interval from diagnosis to surgery (median interval 5 days vs. 22 days; p = 0.09). Of the 15 patients who survived palliative surgery, 11 went on to have repair of PTA (with eight hospital deaths and one late death at conduit replacement), and 4 patients were late deaths after hospital discharge occurring at 29 days, 1.7 months, 1.4 years and 13.3 years after surgery.

Persistent truncus arteriosus repair. A total of 148 of 202 (73%) patients underwent repair of PTA. The proportion of patients who underwent PTA repair increased significantly (p = 0.001) with subsequent birth cohorts as follows: 1953–1967, 4/13 (31%); 1968–1977, 24/41 (59%); 1978–1987, 50/69 (72%) and 1988–1997, 70/80 (88%). Before repair 53/138 (38%) patients were managed in the intensive care unit with 41/138 (30%) patients having been mechanically ventilated for a median of 2 days (range, 1 day to 92 days). Reasons for preoperative ventilation included elective in 15 patients, respiratory compromise in 19, hemodynamic

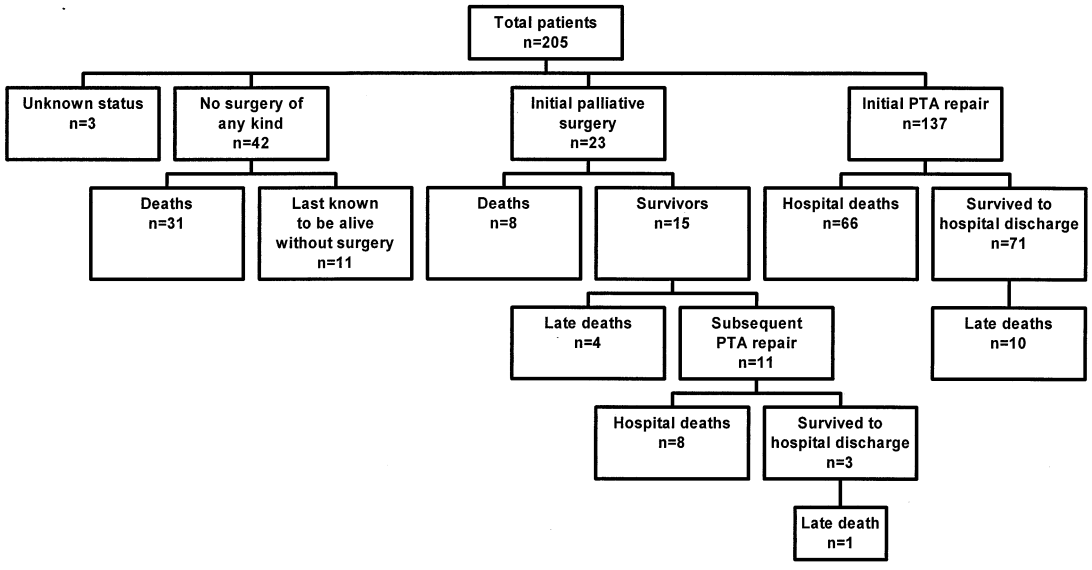

Figure 1. Management and outcomes. PTA = persistent truncus arteriosus.

instability in 4 and unknown reasons in 3 patients. On electrocardiograms obtained just before surgery (n = 134), there was no evidence of myocardial ischemia in 66 patients (49%), ST segment changes only in 30 (23%), T wave flattening or inversion only in 22 (16%), both T wave and ST segment changes in 14 (10%) and T wave changes with a strain pattern in 2 patients (2%).

Median age at repair (n = 147) was 38 days (range, 2 days to 21.1 years) and decreased significantly ($p < 0.001$) with subsequent birth cohorts as follows: 1953–1967, 11.2 years; 1968–1977, 1.1 years; 1978–1987, 1.6 months and 1988–1997, 12 days. Mean cardiopulmonary bypass time (available for 143 patients) was 133 ± 58 min with a mean aortic cross-clamping time (available for 131 patients) of 52 ± 25 min. Circulatory arrest was used in 90 of 132 (68%) patients for which this could be ascertained, with a mean duration (available for 81 of the 90 patients) of 38 ± 20 min. The RV to PA conduit was valved in 144/144 patients, and the type of conduit (n = 144) was a xenograft in 43% (Hancock in 61%, Polystan in 36% and Goretex in 3%), cryopreserved aortic homograft in 37%, cryopreserved pulmonary homograft in 19% and an autograft (direct tissue connection) in 1%. In addition to repair of interrupted aortic arch where applicable at the time of PTA repair, six patients had replacement of the PTA valve with two additional patients having repair of the PTA valve.

Hospital mortality. There were 74 deaths (50%) before hospital discharge, with 43 deaths either in the operating room or within the first 24 h after surgery (range up to 14 days). Cumulative deaths versus consecutive repairs is shown in Figure 2. Causes of death included pulmonary hemorrhage in one patient, intracranial hemorrhage in one and cardiac tamponade in one patient, with the remainder of patients dying from circulatory failure. Hospital mortality improved significantly ($p < 0.001$) with subsequent birth cohorts as follows: 1953–1967, 2/4 (50%); 1968–1977, 15/24 (63%); 1978–1987, 28/50 (56%) and 1988–1997, 29/70 (41%). Since 1995, an aggressive preoperative intensive care management program has been started and combined with one cardiac surgeon designated to repair these patients, with an apparent improvement in hospital mortality during 1996–1997 to 2 of 11 (18%).

Factors associated with hospital mortality with PTA repair were sought (Table 2). Hospital mortality was not significantly related to preoperative PTA anatomy (PA connections), PTA valve stenosis or regurgitation, number of PTA valve cusps, presence of a major associated cardiac anomaly, previous palliative procedures, preoperative management in the intensive care unit or use of mechanical ventilation, presence of ischemic changes on the preoperative electrocardiogram, age at PTA repair or cardiopulmonary bypass time. Increased hospital mortality was significantly related to earlier date of surgical repair in the overall experience.

Total early mortality. If deaths occurring before PTA repair are combined with hospital mortality after repair, then total early mortality was 113/202 (56%). Total early mortality did not change significantly ($p = 0.20$) with subsequent birth cohorts as follows: 1953–1967, 5/12 (42%); 1968–1977, 28/41 (68%); 1978–1987, 42/69 (61%) and 1988–1997, 38/80 (48%), with a mortality in the last 5 years of 24/41 (59%) and in the most recent 2 years of 4/11 (36%).

Late mortality. There were 11 late deaths (15%) in the 74 hospital survivors after PTA repair, which occurred at a median interval after repair (n = 10) of 5.5 months (range, 1.8 months to 5.3 years). Cause of late death was related to

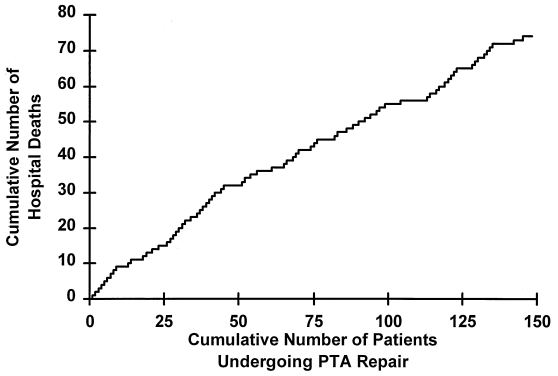

Figure 2. Cumulative hospital mortality with consecutive surgical repairs of persistent truncus arteriosus. PTA = persistent truncus arteriosus.

pulmonary infection in three patients, pulmonary hypertension in one, cardiac failure in one, arrhythmia in one, thrombosis of the conduit valve in one, hospital death after conduit replacement in one and was unknown in three patients. Kaplan Meier survival estimates after hospital discharge in survivors after repair are shown in Figure 3, with survival of 83% up to 20.3 years. Survival after hospital discharge was not significantly related to demographics, PTA and PTA valve anatomy, previous palliative procedures, age and date of PTA repair or cardiopulmonary bypass time.

Subsequent procedures. Persistent truncus arteriosus valve replacement was required in four patients, with a second valve replacement required in one of these patients. Conduit replacement was required during follow-up procedures in 34 of the 74 survivors (46%), with Kaplan-Meier estimates of freedom from initial conduit replacement shown in Figure 4. A second conduit replacement was performed in seven patients with one patient requiring a third replacement. Freedom from initial conduit replacement was not significantly related to patient age or date of PTA repair but was significantly increased with increasing size of initial conduit placed (log rank test $p = 0.02$; Wilcoxon test $p = 0.01$). This was also significantly related to the type of initial homograft material (log rank test $p = 0.002$, Wilcoxon test $p = 0.008$), with pulmonary homografts lasting the longest, aortic homografts next followed by xenografts (Fig. 5). Interventional cardiac catheterization was performed in 31 patients (42%), initially for branch PA stenoses in 19 patients, for conduit or conduit valve stenosis in 9 and for both in 4 patients, with 1 patient having balloon dilation of aortic arch obstruction. Subsequent additional interventional catheterization procedures were performed in 13 of these patients. The use of interventional cardiac catheterization to address conduit and conduit valve stenosis ($n = 13$ patients) before first conduit replacement or last known follow-up procedure did not significantly relate to freedom from conduit replacement (log rank test $p = 0.93$; Wilcoxon test $p = 0.96$) versus those patients who did not have an interventional procedure. However, if an assumption is made that the date of the interventional procedure might be the date of initial conduit replacement if interventional catheterization had not been performed, then these patients would have been significantly more likely to have earlier initial conduit replacement (log rank test $p < 0.001$; Wilcoxon test $p < 0.001$) (Fig. 6).

DISCUSSION

Natural history. The natural history of PTA in the presurgical era has not been well-documented. In 1957, Anderson et al. (7) described a group of 14 patients with PTA, with 11 of them diagnosed at autopsy and 3 confirmed at surgery. The median age at diagnosis was 3 months. In 1976, Calder et al. (8) noted that the diagnosis in their study group of 100 patients was still primarily made at autopsy (79%) at a median age of 5 weeks. Most patients died in early infancy (7–10). In our study, we found that 31 of 42 patients (74%) that had no surgery died at a median age of 11 days with a range from birth to 19.4 years, with 84% (26/31) being younger than 6 months old. Hicken et al. (9) first reported a case of a woman with PTA that died at the age of 38 years. Three published long-term survival studies of unrepaired patients report mortality rates of 61% with death occurring at a median age of 1.5 years (10), 81% with a median age of 5 weeks (8) and 100% with a median age of 4.5 months (7), respectively.

Palliative surgical procedures. The role of palliative surgical procedures such as PA banding to manage PTA has changed considerably during the years. Trends toward decreased palliative procedures were accompanied by trends toward younger age at PTA repair. In the past, PTA patients underwent PA banding if medical treatment failed to control congestive heart failure (5), often with a view to PTA repair at an older age. Palliation with PA banding is currently not believed to have any advantages (8,11) and is associated with high mortality rates (4,5,12–16). Poirier et al. (16) reported in a literature review regarding PA banding in PTA patients that the pooled overall mortality rate was 59% (45/76) and ranged between individual series from 0% (0/6) to 100% (3/3). Singh et al. (15) reported 11 deaths (73%) after PA banding in 15 patients. These reports of high mortality are similar to our experience, where 11/23 (48%) patients either died early or late after PA banding. Furthermore, PA banding does not sufficiently prevent pulmonary vascular obstructive disease, which has been reported to occur in 10% to 15% of patients undergoing PA banding (5); this complication renders these patients inoperable. Pulmonary artery banding is now reserved for patients that have increased pulmonary blood flow and are not deemed suitable candidates for definitive repair of their lesion. The decreasing use of initial palliative procedures in our series from 25% in 1953–1967 to 2% in 1988–1997 reflects these changes in management policy.

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Table 2. Factors Associated with Death Before Hospital Discharge in Patients Undergoing Repair of Persistent Truncus Arteriosus (N = 148)

Variable	Hospital Mortality	%	p*
PTA anatomy			
PA connection			
Single common pulmonary trunk	50/96	52	
Branch PA's from separate orifaces but close together on PTA	16/35	46	
Other	8/15	53	0.80
Number of PTA valve cusps			
Two	4/7	57	
Three	36/69	52	
Four or more	24/44	50	0.95
Degree of PTA valve stenosis			
None	25/52	48	
Mild	20/42	48	
Moderate to severe	6/10	60	0.77
Degree of PTA valve regurgitation			
None	20/42	48	
Mild	38/77	49	
Moderate to severe	8/14	57	0.83
Associated major cardiovascular anomaly			
Absent	64/133	48	
Present	10/13	67	0.28
Previous palliative surgical procedure			
Absent	64/133	48	
Present	8/11	73	0.21
Preoperative management in the intensive care unit			
Absent	43/85	51	
Present	28/53	53	0.80
Preoperative mechanical ventilation			
Absent	49/97	51	
Present	22/41	54	0.74
Presence of preoperative ischemic changes on electrocardiogram			
Absent	31/67	46	
Present	38/67	57	0.23

Variable	Hospital Survivors (n = 74)	Deaths (n = 74)	p
Median (range) date of surgery	04/1989 (09/1979 to 09/1997)	09/1986 (04/1971 to 06/1997)	0.02
Median (range) age at repair	58 days (n = 73, 2 days to 21.1 yrs)	23 days (2 days to 9.6 yrs)	0.19
Mean (±1 SD) cardiopulmonary bypass time (min)	131 ± 57 (n = 70)	136 ± 61 (n = 73)	0.63

PA = pulmonary artery; PTA = persistent truncus arteriosus; SD = standard deviation; yrs = years.
*From chi-square tests to compare categorical variables, Kruskal-Wallis analysis of variance to compare medians and Student *t* test to compare means.

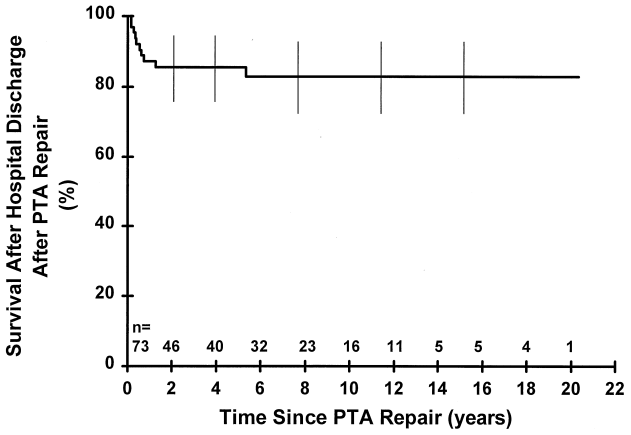

Figure 3. Kaplan-Meier estimates of late death after hospital discharge in survivors of repair of persistent truncus arteriosus. **Vertical lines** represent 95% confidence limits. PTA = persistent truncus arteriosus.

Age at repair. Age at PTA repair has been decreasing over the years. Overall median age at repair in this study was 38 days, but decreased significantly with subsequent birth cohorts from 11.2 years to 12 days. This trend is supported in the published literature from the past 30 years, with reported median ages ranging from 3.5 months to 7.3 years (1,3,17–19). More recent studies published in the last 15 years report that age at repair has decreased dramatically, with median ages ranging from 8 to 41 days (11,20–23). A possible explanation for this trend is the improvements in neonatal cardiothoracic surgery combined with a desire to prevent pulmonary vascular obstructive disease in the patients who are at very high risk.

Hospital mortality after PTA repair. Early mortality rates after PTA repair vary considerably in previous series. Sano et al. (23) reported no early mortality in a series of seven patients undergoing repair from 1985–1989. Other series from a similar time period report early mortality rates of 9% (1986–1988) (11), 11% (1986–1992) (21), 17.5% (1986–1991) (22) and 15.6% (1982–1990) (4), although in the latter study, the mortality rate decreased to 8.3% during the last 4 years. Series before 1982 have shown higher mortality rates, varying from 11% (12) to 42% (13), with most series having early mortality rates of more than 25% (13,14,18,19,24). This trend toward decreasing mortality with time was observed in our large series. Hospital mortality improved significantly with subsequent birth cohorts, although it still remained high—41% for the last birth cohort (1988–1997). During 1996–1997, however, hospital mortality further declined to 18%, which is comparable with contemporary reports.

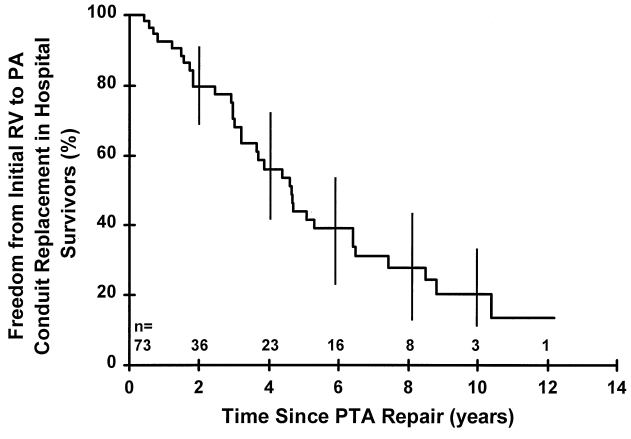

Figure 4. Kaplan-Meier estimates of freedom from first right ventricle to pulmonary artery conduit replacement after repair of persistent truncus arteriosus. Data relates to hospital survivors after repair of persistent truncus arteriosus; **vertical lines** represent 95% confidence limits. PA = pulmonary artery; PTA = persistent truncus arteriosus; RV = right ventricle.

Factors associated with mortality after PTA repair. We noted only one significant factor associated with increased mortality before hospital discharge after PTA repair—earlier date of surgical repair in the overall experience. Age at repair was not significantly related to hospital mortality. Although these findings are comparable with those of some studies (3,17,21), Lacour-Gayet et al. (20) have reported that age younger than one month remains an incremental risk factor. Others have reported that increasing age was a risk factor for hospital mortality (18,22). Previous palliative surgeries performed on patients that subsequently went on to have PTA repair did not significantly increase hospital mortality. However, it must be mentioned that in our experience, 8 (73%) of the 11 patients that had repair after a palliative procedure did not survive to hospital discharge.

The presence of interruption of the aortic arch has been previously reported to be a risk factor for mortality. Pearl et al. (4) reported that the presence of interruption of the aortic arch increased early mortality with PTA repair. Bove et al. (21) reported that associated cardiac defects were not significantly associated with increased mortality. We likewise did not identify an association between other cardiac defects and hospital mortality. This is in contrast with the series reported by Hanley et al. (22) in which interruption of the aortic arch was a significant risk factor for mortality after PTA repair.

Conduit reoperations. Thirty-four patients underwent 42 conduit replacements. There was one hospital death after replacement. Smaller conduit size was a significant factor associated with a shorter time to initial conduit reoperation. Use of a pulmonary homograft versus an aortic homograft or a xenograft was associated with greater initial conduit longevity. Heinemann et al. (25) noted similar results. They noted that the use of aortic homografts are a significant risk factor for early replacement and encouraged the use of a pulmonary homograft. There are many advantages in using a pulmonary homograft (26). However, Morshuis et al. (27) reported the use of a pulmonary homograft in the reconstruction of PTA in an infant, with severe conduit valve

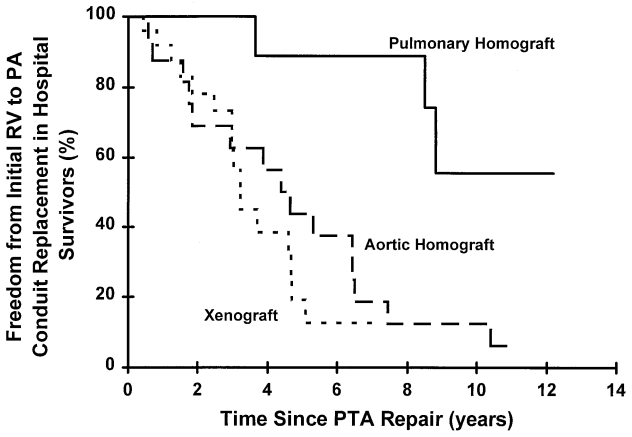

Figure 5. Kaplan-Meier estimates of freedom from initial conduit replacement related to the type of conduit. Data relates to hospital survivors after repair of persistent truncus arteriosus. Log rank p = 0.002; Wilcoxon test p = 0.008. PA = pulmonary artery; PTA = persistent truncus arteriosus; RV = right ventricle.

stenosis one year later. They felt that prevention of pulmonary regurgitation was the only advantage to the valved conduit. Interventional cardiac catheterization (balloon dilation with or without endovascular stenting) to address conduit and conduit valve stenosis was performed in 31 patients before the first conduit reoperation. This did not appear to have a significant influence on freedom from conduit replacement, unless the assumption was made that conduit reoperation would have occurred at the time of

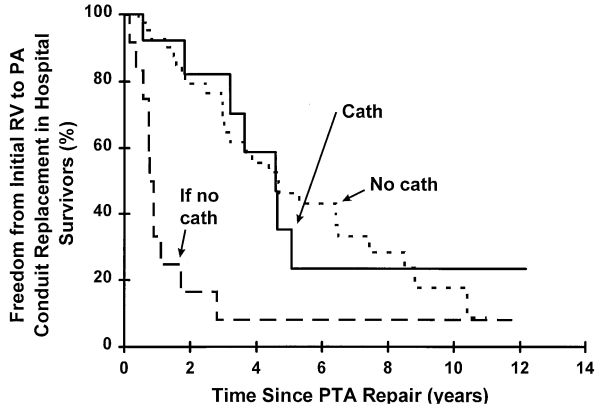

Figure 6. Kaplan-Meier estimates of freedom from initial conduit replacement related to the influence of transcatheter balloon dilation with or without endovascular stenting of conduit or conduit valve stenosis. Data relates to hospital survivors following repair of persistent truncus arteriosus. **Dashed line** represents patients who did not have any intervening interventional cardiac catheterization to address conduit obstruction (No cath); **solid line** represents patients who had intervening interventional catheterization (Cath); and **broken line** represents the same patients as the solid line but under the assumption that if interventional catheterization were not performed they would have had conduit replacement at that date (If no cath). PA = pulmonary artery; PTA = persistent truncus arteriosus; RV = right ventricle.

catheter intervention if it had not been performed. This compares favorably with published studies of balloon dilations and stent implantations in RV to PA conduits. Lloyd et al. (28) reported successful dilation of stenotic conduit valve in three of six patients, with important increases in conduit longevity. Ensing et al. (29) reported relief of conduit obstruction in 8 of 11 patients undergoing balloon dilation. One patient returned for conduit replacement 1.5 years after the procedure. Powell et al. (30) described 44 patients who underwent endovascular stent implantation with important improvements in conduit longevity. However, Zeevi et al. (31) reported less positive outcomes, with successful relief of obstruction in three of nine patients, one of whom developed recurrence and required conduit replacement within 1 year after the procedure.

PTA valve replacement. Insufficiency of the truncal valve before initial PTA repair has been associated with a higher risk of early mortality (4,12,18,22) and poorer long-term survival (3,18,19). Therefore, it is believed that replacement or repair of a significantly regurgitant or stenotic truncal valve at the time of PTA repair may be warranted (4,12,18,19,21,22,32). Bove et al. (11) have asserted that the truncal valve should be left alone at the time of PTA when the degree of regurgitation is less than severe. Other series' researchers have noted that preoperative truncal valve regurgitation was not significantly associated with increased early mortality after PTA repair (3,17,18,20,21,24). Bove et al. (21) and Marcelletti et al. (19) noted similar results on early mortality, but assert that significant truncal valve regurgitation complicates the operative and the early postoperative course. Our study, similarly, did not show a significant relation between hospital mortality after PTA repair and the preoperative degree of truncal valve regurgitation.

As patients with important truncal valve dysfunction often undergo truncal valve repair or replacement, controversy exists as to whether the observed increased mortality at PTA repair is associated with valve repair or replacement or the presence of prerepair regurgitation. In our series, truncal valve replacement at time of repair was performed for six patients—three of whom had mild truncal valve regurgitation (one hospital death, and one late death 9 months after surgery), one moderate and two with severe regurgitation (both died before hospital discharge). This represents a total mortality rate of 67%. Brizard et al. (17) reported that mortality related to replacement or repair of the truncal valve at repair may explain the finding of truncal valve regurgitation as a risk factor for hospital mortality. McElhinney et al. (14) also report a high mortality after truncal valve replacement. In their series of seven patients who had replacement of their valve, 5 died early and 2 patients died late after the operation. One of the late deaths occurred 8 months after initial truncal valve replacement and two months after a second valve replacement was performed. All but 1 of the 7 patients had severe truncal valve regurgitation preoperatively. In our series, 5 truncal valve replacements

were performed in 4 patients during follow-up procedures after PTA repair, 2 of whom had mild and 2 of whom had moderate regurgitation before repair. None of these patients died. Unfortunately, the number is too small to draw conclusions, but prerepair truncal valve regurgitation may be a risk factor for need for late truncal valve replacement, although this may be dependent on the degree of prerepair regurgitation (17). Freedom from valve replacement is better in those with mild regurgitation than in those with moderate or severe regurgitation (14).

Conclusions. We conclude that there have been significant trends toward improved mortality into the current era, which have been marked by concomitant trends towards improved early diagnosis and primary neonatal repair. Nonetheless, important preoperative morbidity and mortality continue to occur. Hospital mortality after repair, although improved, remains significant and ongoing morbidity related to conduit obstruction and reoperation, and truncal valve dysfunction and replacement or repair continues to dominate long term outcomes. There is growing evidence to support the use of interventional cardiac catheterization to address conduit and conduit valve obstructions and improve conduit longevity.

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REFERENCES

1. Slavik Z, Keeton BR, Salmon AP, Sutherland GR, Fong LV, Monro JL. Persistent truncus arteriosus operated during infancy: long-term follow-up. Ped Card 1994;15:112–5.
2. Williams RL, Sommerville R. Truncus arteriosus. In: Garson A, Bricker JT, McNamara DG, editors. The Science and Practice of Pediatric Cardiology. Philadelphia, PA: Lea and Febiger, 1990:1127–33.
3. Rajasinghe HA, McElhinney DB, Reddy VM, Mora BN, Hanley FL. Long-term follow-up of truncus arteriosus repaired in infancy: a twenty-year experience. J Thorac Cardiovasc Surg 1997;113:869–79.
4. Pearl JM, Laks H, Drinkwater DC, et al. Repair of truncus arteriosus in infancy. Ann Thorac Surg 1991;52:780–6.
5. Sharma AK, Brawn WJ, Mee RBB. Truncus arteriosus: surgical approach. J Thorac Cardiovasc Surg 1985;90:45–9.
6. McGoon DC, Rastelli GC, Ongley PA. An operation for the correction of truncus arteriosus. JAMA 1968;205:69–73.
7. Anderson RC, Obata W, Lillehei W. Truncus arteriosus: clinical study of 14 cases. Circulation 1957;16:586–98.
8. Calder L, Van Praagh R, Van Praagh S, et al. Truncus arteriosus communis: clinical, angiocardiographic and pathologic findings in 100 patients. Am Heart J 1976;92:23–38.
9. Hicken P, Evans D, Heath D. Persistent truncus arteriosus with survival to the age of 38 years. Br Heart J 1966;28:284–6.
10. Marcelletti C, McGoon DC, Mair DD. The natural history of truncus arteriosus. Circulation 1976;54:108–11.
11. Bove EL, Beekman RH, Snider AR, et al. Repair of truncus arteriosus in the neonate and young infant. Ann Thorac Surg 1989;47:499–505.
12. Ebert PA, Turley K, Stanger P, Hoffman JIE, Heymann MA, Rudolph AM. Surgical treatment of truncus arteriosus in the first 6 months of life. Ann Surg 1984;200:451–6.
13. Stark J, Candhi D, de Leval M, Macartney F, Taylor JFN. Surgical treatment of persistent truncus arteriosus in the first year of life. Br Heart J 1978;40:1280–7.
14. McElhinney DB, Reddy VM, Rajasinghe HA, Mora BN, Silverman NH, Hanley FL. Trends in the management of truncal valve insufficiency. Ann Thorac Surg 1998;65:517–24.
15. Singh AK, De Leval MR, Pincott JR, Stark J. Pulmonary artery banding for truncus arteriosus in the first year of life. Circulation 1974;49:III17–9.
16. Poirier RA, Berman MA, Stansel HC Jr. Current status of the surgical treatment of truncus arteriosus. J Thorac Cardiovasc Surg 1975;69:169–82.
17. Brizard CP, Cochrane A, Austin C, Nomura F, Karl TR. Management strategy and long term outcome for truncus arteriosus. Eur J Cardio Thorac Surg 1997;11:687–96.
18. Di Donato RM, Fyfe DA, Puga FJ, et al. Fifteen-year experience with surgical repair of truncus arteriosus. J Thorac Cardiovasc Surg 1985;89:414–22.
19. Marcelletti C, McGoon DC, Danielson GK, Wallace RB, Mair DD. Early and late results of surgical repair of truncus arteriosus. Circulation 1977;55:636–41.
20. Lacour-Gayet F, Serraf A, Komiya T, et al. Truncus arteriosus repair: influence of techniques of right ventricular outflow tract reconstruction. J Thorac Cardiovasc Surg 1996;111:849–56.
21. Bove EL, Lupinetti FM, Pridjian AK, et al. Results of a policy of primary repair of truncus arteriosus in the neonate. J Thorac Cardiovasc Surg 1993;105:1057–66.
22. Hanley FL, Heinemann M, Jonas RA, et al. Repair of truncus arteriosus in the neonate. J Thorac Cardiovasc Surg 1993;105:1047–56.
23. Sano S, Brawn WJ, Mee RB. Repair of truncus arteriosus and interrupted aortic arch. J Card Surg 1990;5:157–62.
24. Spicer RL, Behrendt D, Crowley DC, et al. Repair of truncus arteriosus in neonates with the use of valveless conduit. Circulation 1984;70:I26–9.
25. Heinemann MK, Hanley FL, Fenton KN, Jonas RA, Mayer JE, Castaneda AR. Fate of small homograft conduits after early repair of truncus arteriosus. Ann Thorac Surg 1993;55:1409–12.
26. Livi U, Kay P, Ross D. The pulmonary homograft: an improved conduit for RVOT reconstruction. Circulation 1986;74:II250.
27. Morshuis WJ, van de Wal HJ, Tanke RJ, Lacquet LK. Reconstruction of the right ventricular outflow tract in truncus arteriosus types I and II with pulmonary homograft. J Thorac Cardiovasc Surg 1992;104:842–3.
28. Lloyd TR, Marvin WJ, Mahoney LT, Lauer RM. Balloon dilation valvuloplasty of bioprosthetic valves in extracardiac conduits. Am Heart J 1987;114:268–74.
29. Ensing GJ, Hagler DJ, Seward JB, Julsrud PR, Mair DD. Caveats of balloon dilation of conduits and conduit valves. J Am Coll Cardiol 1989;14:397–400.
30. Powell AJ, Lock JE, Keane JF, Perry SB. Prolongation of RV-PA conduit life span by percutaneous stent implantation. Intermediate-term results. Circulation 1995;92:3282–8.
31. Zeevi B, Keane JF, Perry SB, Lock JE. Balloon dilation of postoperative right ventricular outflow obstructions. J Am Coll Cardiol 1989;14:401–8.
32. Black MD, Adatia I, Freedom RM. Truncal valve repair: initial experience in neonates. Ann Thorac Surg 1998;65:1737–40.